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CONGENITAL HEPATIC FIBROSIS WITH POLYCYSTIC KIDNEY DISEASE PRESENTED AS PORTAL HYPERTENSION IN A 7 YEAR OLD FEMALE CHILD – A RARE CASE REPORT

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INTRODUCTION

Congenital hepatic fibrosis (CHF) is a rare, autosomal recessive disorder, clinically characterized by hepatic fibrosis and portal hypertension. CHF results from ductal plate malformation (DPM) of the intrahepatic bile ducts. Four clinical forms can be observed: portal hypertensive, cholangitic, mixed and latent. CHF is one of the "fibropolycystic diseases" which also include several conditions with a variety of intrahepatic bile duct dilatation and associated periportal fibrosis such as Caroli disease, autosomal recessive and dom-inant polycystic kidney disease (ARPKD or ADPKD), Ivemark, Jeune, Joubert, Bardet-Biedl, Meckel-Gruber and Arima syndromes. Most of them are accompanied by progressive cystic degeneration of the kidneys. We present the case of a 7-year-old female patient with CHF with POLYCYSTIC KIDNEY DISEASE and a review of the literature.

CASE DESCRIPTION

We present the case of a 7-year-old female patient with CHF, with nonspecific clinical manifestation, which made it very difficult to diagnose this disease.

A previously healthy 7-year-old girl was referred to the hospital by her pediatrician with an asymp-tomatic splenomegaly which was revealed in a physi-cal examination (4 cm below the left costal margin). The enlarged spleen was confirmed by abdominal ultra sonography (65 mm × 155 mm). Hepatotropic virus infection, cytomegalovi-rus, Epstein-Barr virus, and thrombophilia were ex-cluded. Computed tomography (CT) imaging showed spleen enlargement (150 mm × 100 mm), normal liver size and also the presence of portosystemic collateral vessels along the lesser curvature of the stomach, car-diac region, body of the pancreas, in the porta hepatis area and in the splenic hilum.

The differential diagnosis excluded Wilson's disease and α-1-anti-trypsin deficiency. Gastroscopy visualized a low-grade esophageal varices (with trophic changes) located in the lower third and features of portal hypertensive gastropathy. There were no indications to perform en-doscopic variceal ligation (EVL). The ultrasonography and CT scan excluded prehepatic flow block. Kidneys-Rt. And Lt. – B/L Grade I echogenicity with multiple tiny cysts upto 8mm noted in both kidney and confirmed normal liver function. The patient was qualified for a percutaneous liver biopsy.

A histopathological examination of the liver biopsy showed portal and periportal fibrosis with the pres-ence of fibrous bands between the portal tracts without signs of cirrhosis (Fig. 3)

Classically, CHF is associated with ARPKD, and some authors believe that CHF and ARPKD represent a single disorder, with a wide spectrum of manifesta-tions. Others argue that there are two distinct disor-ders with similar hepatic and biliary lesions.

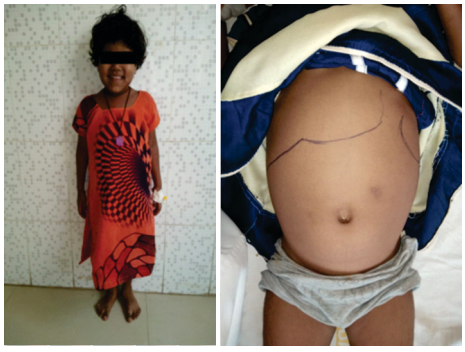

fig 1.and 2. child presented with hepato spleeno megaly

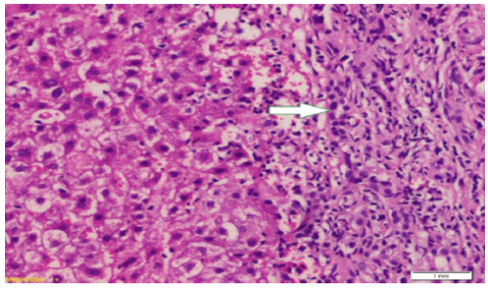

fig 3.histology shows portal and peri portal fibrosis

CONCLUSIONS

1. Congenital hepatic fibrosis (CHF) is a rare, autoso-mal recessive disorder, clinically characterized by hepatic fibrosis and portal hypertension.
2. A liver biopsy usually confirms the diagnosis and may distinguish between portal hypertensive, chol-angitic and mixed hypertensive-cholangitic type.